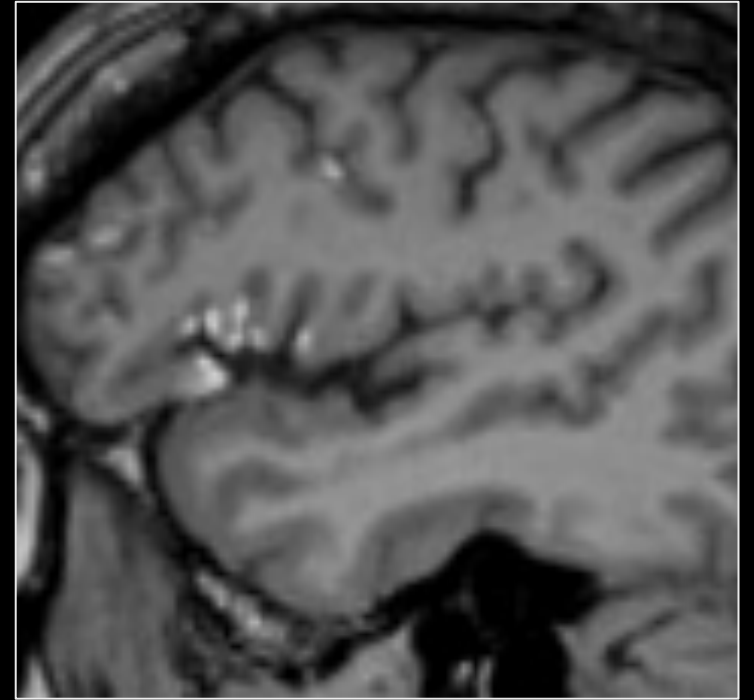
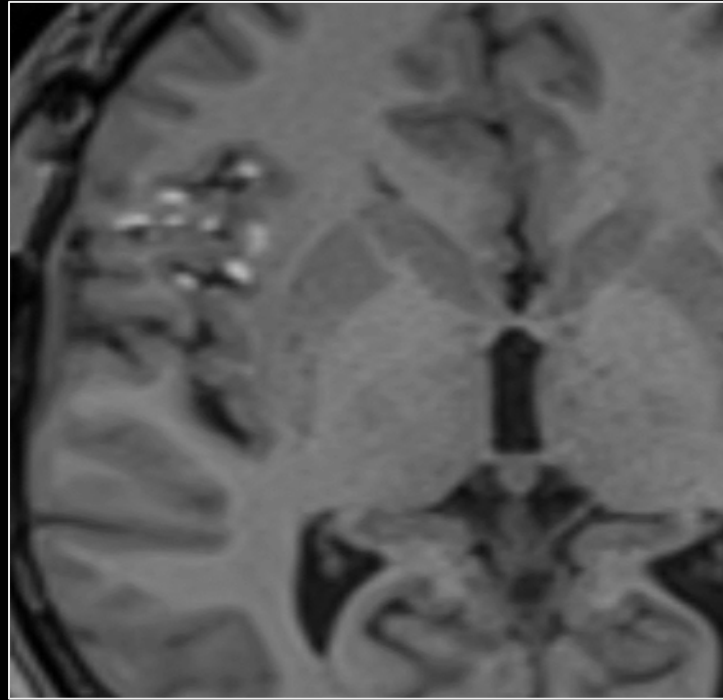
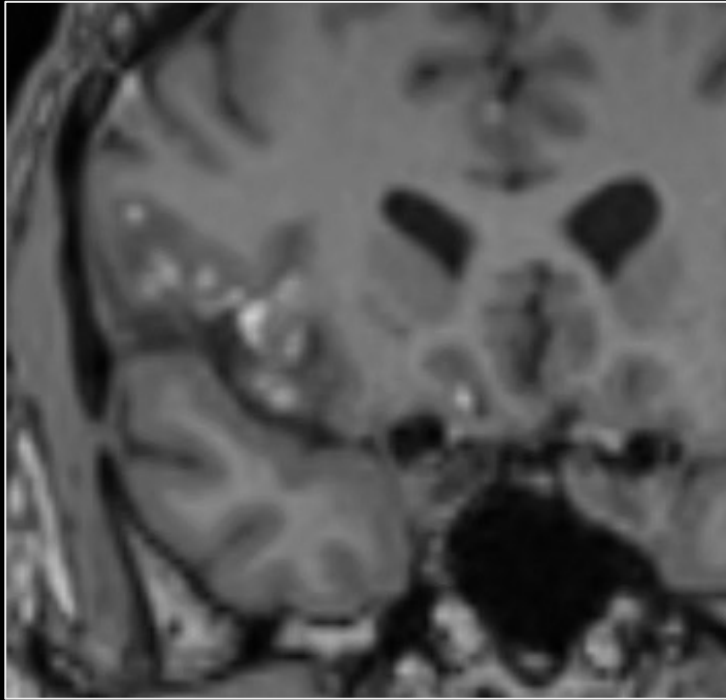


# A Rare Case of Leptomeningeal Melanocytosis



**Gibson Klaphthor, MD**  
**Paul M. Bunch, MD**



# Clinical Presentation

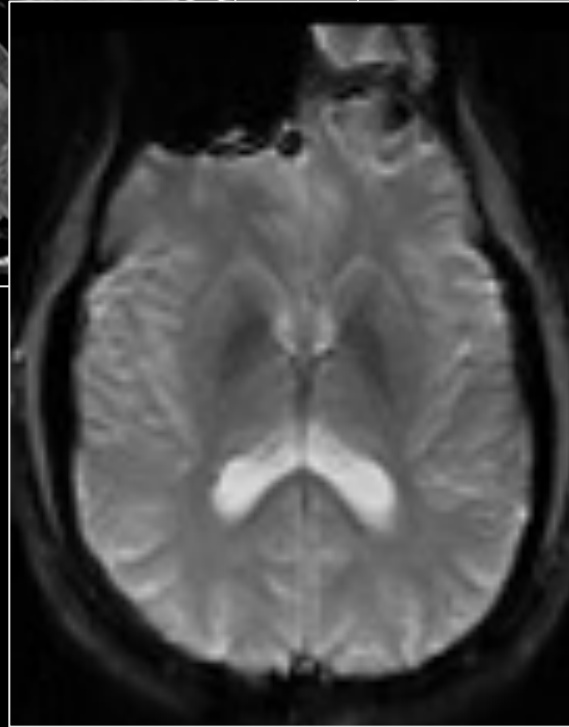
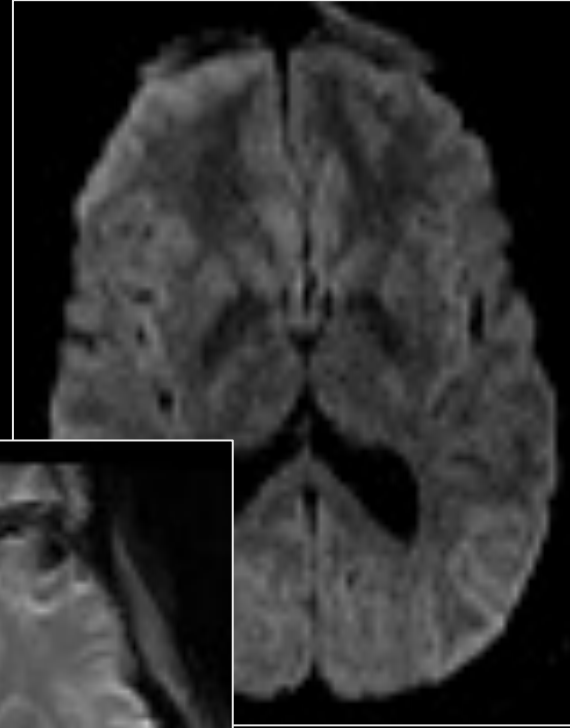
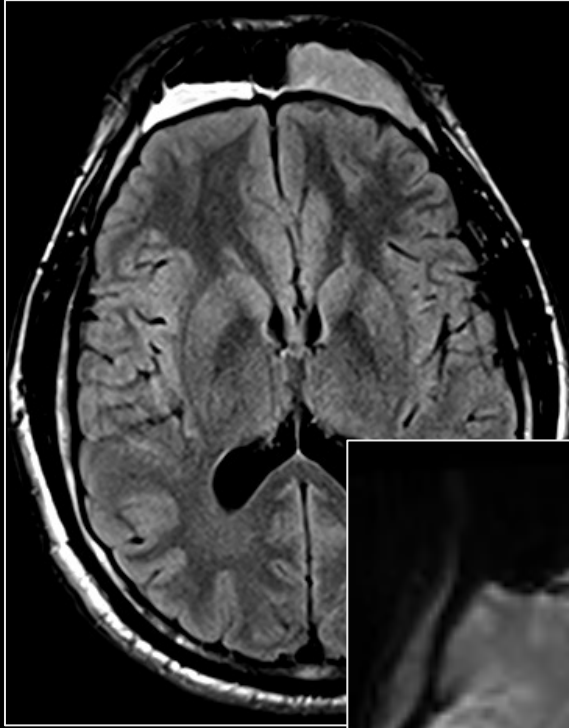
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A 34-year-old male with history of thyroid carcinoma presented with multiple episodes of headaches with left-sided weakness and tingling.

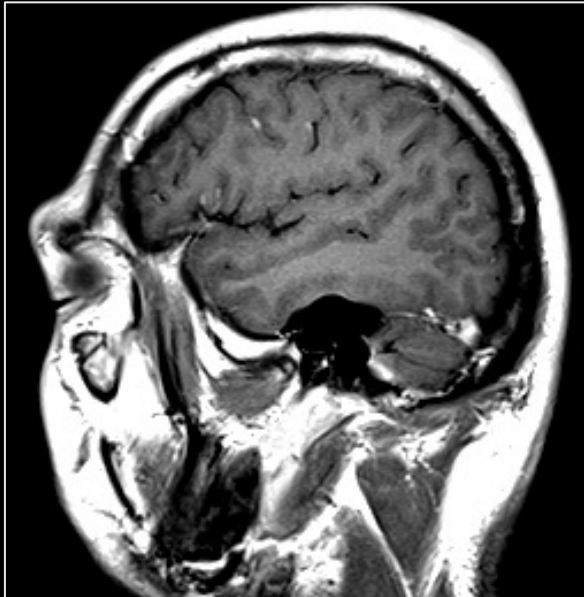
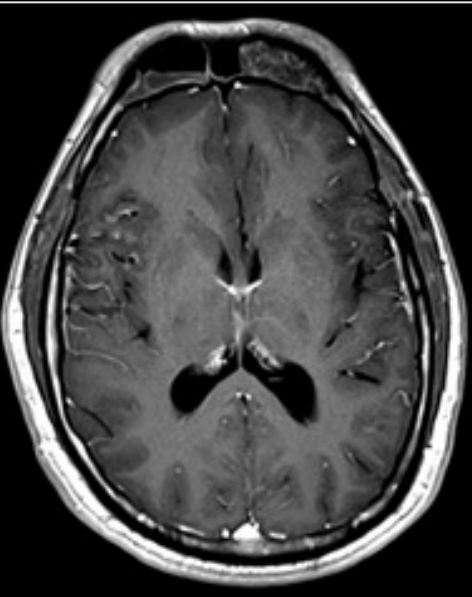
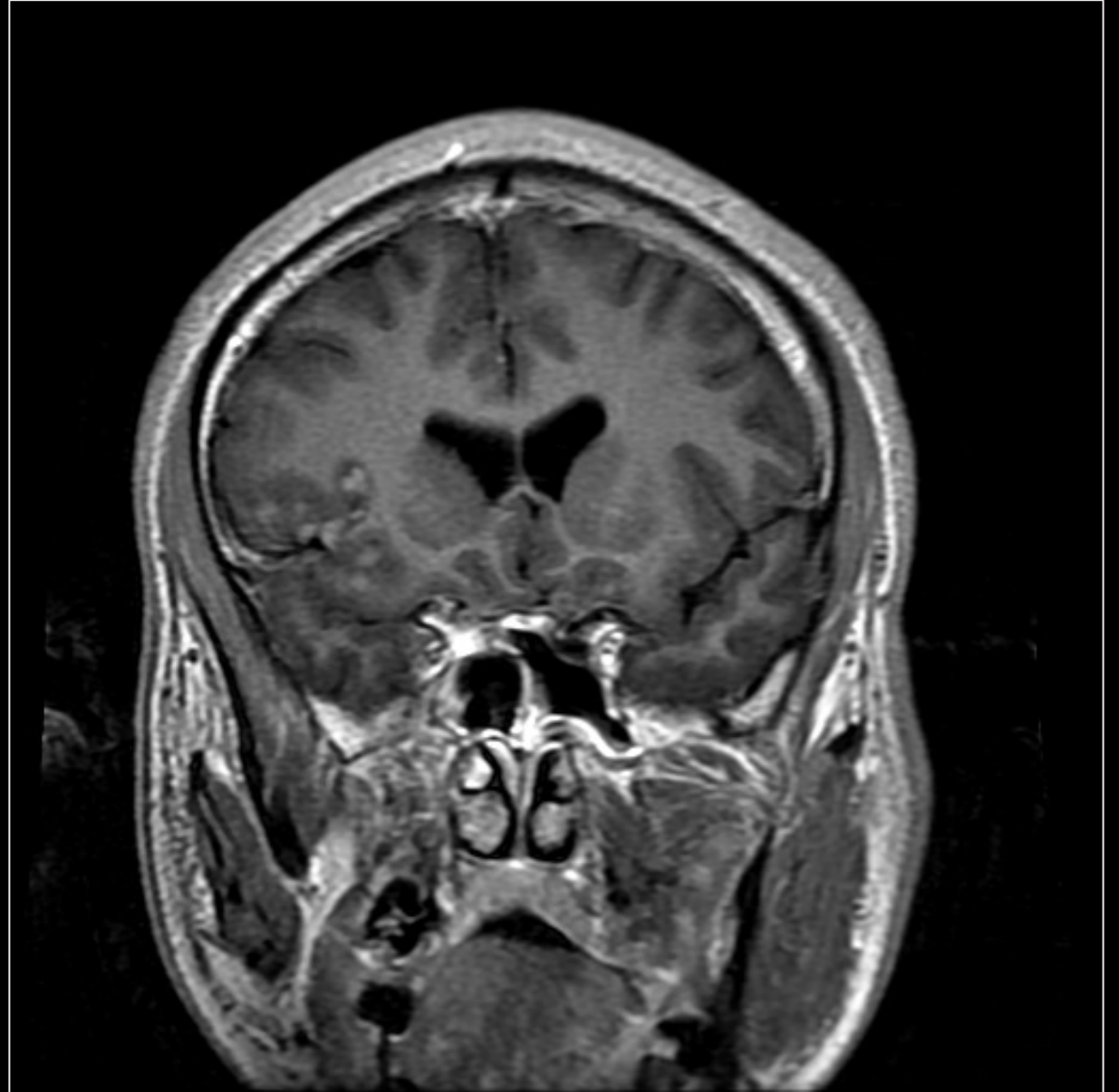
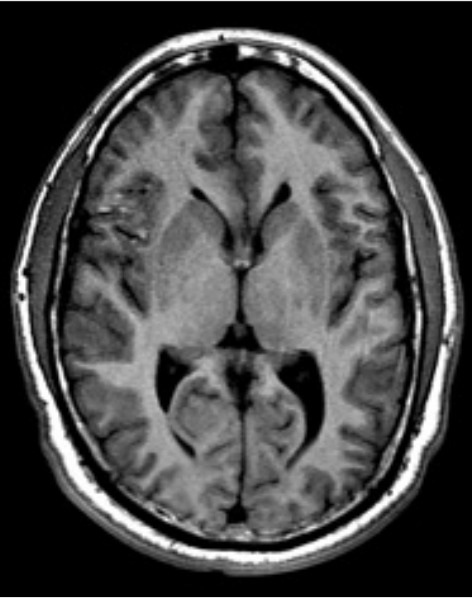
# Imaging



# Imaging

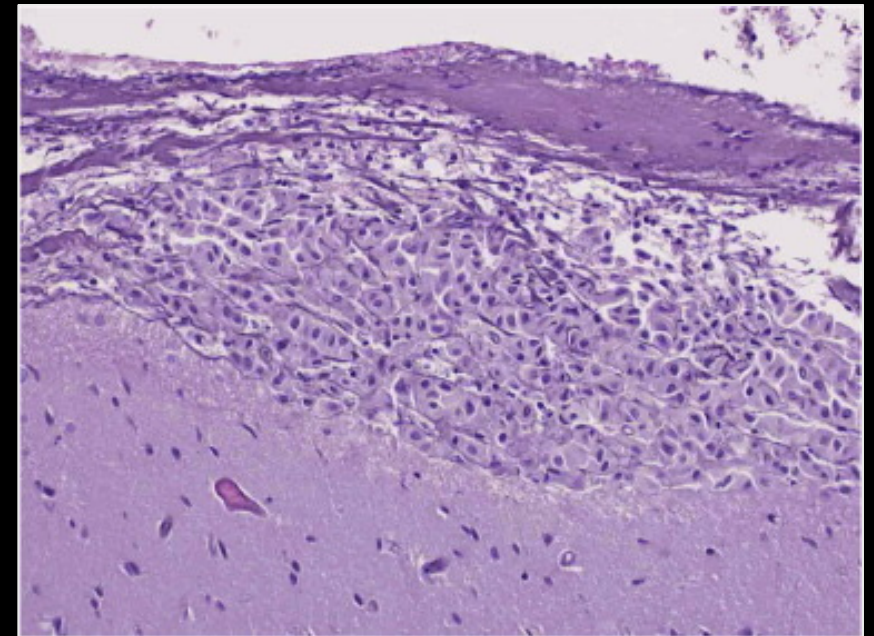
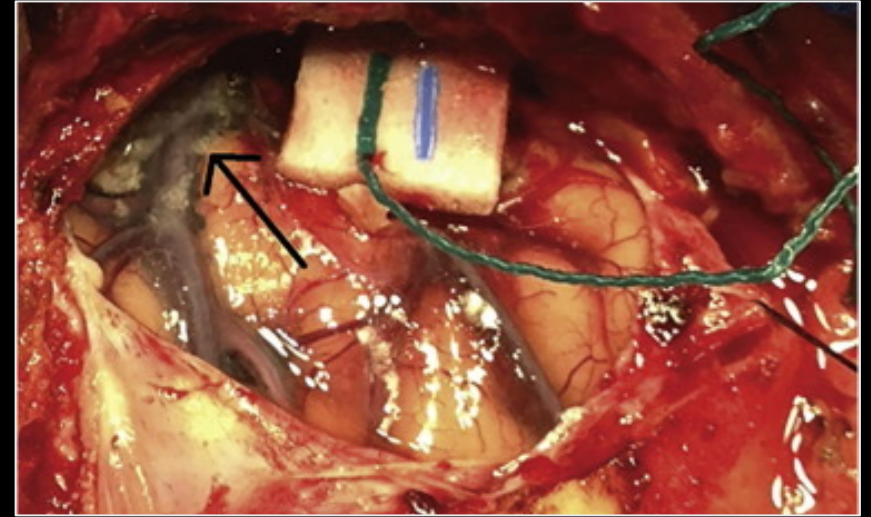


# Imaging



# Diagnosis

- Right frontal craniotomy for biopsy.
- Histopathologic evaluation showed leptomeningeal melanocytosis.



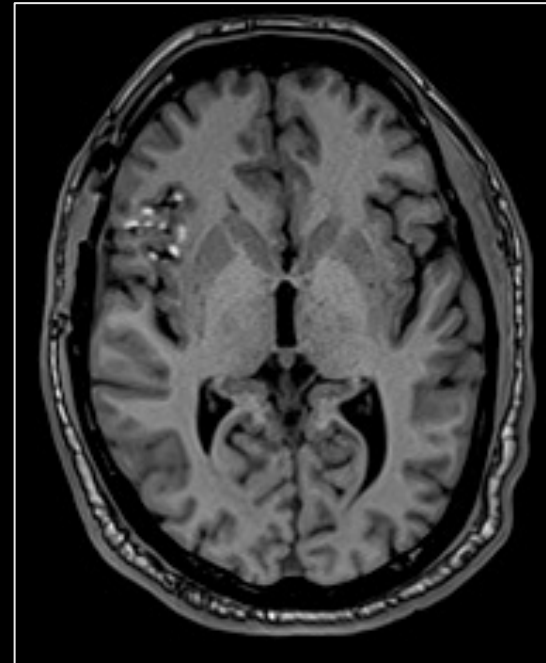
# Management

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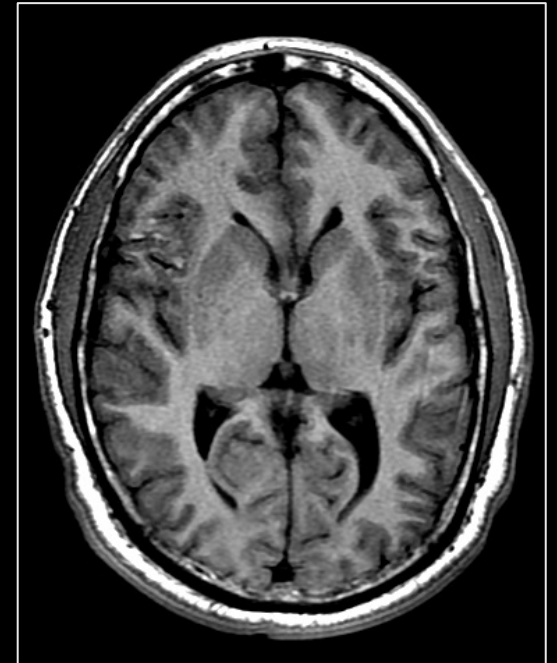
- Treated with solumedrol, cyclophosphamide, and levetiracetam.
- Cyclophosphamide discontinued due to development of hemorrhagic cystitis.
- Serial MR imaging follow-up.

# Outcome

- No MRI evidence of disease progression over >10 years.
- Seizures well-controlled on levetiracetam.
- Patient doing well clinically.



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# Take Home Points

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- Primary leptomeningeal melanocytosis is a **rare** (~1 in 10 to 20 million) benign CNS tumor of neural crest origin believed to arise from melanocyte precursors (melanoblasts).
- Variable clinical symptoms (headache, neck pain, focal neurologic deficits, seizures, symptoms of increased intracranial pressure)
- Key imaging finding is **intrinsically T1 hyperintense lesions** with or without enhancement; typically no associated edema.
- Because leptomeningeal melanoma metastases are far more common, patients **must be evaluated for dermatologic and ocular melanoma.**
- Biopsy required for definitive diagnosis. Treatment is controversial.

# Additional Reading

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- Honigberg MC, et al. Primary leptomeningeal melanocytosis presenting as chronic meningitis. *J Clin Neurosci*, 2014; 21(6):1056-1058.
- Küsters-Vandeveldde HVN, et al. Primary melanocytic tumors of the central nervous system: a review with focus on molecular aspects. *Brain Pathol*, 2015; 25(2):209-226.
- Hossain FA, et al. CT and MRI findings in leptomeningeal melanocytosis. *Radiol Case Rep*, 2019; 18;15(3):186-189.
- Selvarajan JMP, et al. Pearls & Oy-sters: Primary diffuse leptomeningeal melanocytosis: a diagnostic conundrum. *Neurology*, 2023; 1;101(5):e576-e580.